**Medical Policy Manual** Surgery, Policy No. 159

# *Vertical Expandable Prosthetic Titanium Rib*

**Effective**: July 1, 2018

**Next Review:** May 2019
**Last Review:** May 2018

## IMPORTANT REMINDER

Medical Policies are developed to provide guidance for members and providers regarding coverage in accordance with contract terms. Benefit determinations are based in all cases on the applicable contract language. To the extent there may be any conflict between the Medical Policy and contract language, the contract language takes precedence.

PLEASE NOTE: Contracts exclude from coverage, among other things, services or procedures that are considered investigational or cosmetic. Providers may bill members for services or procedures that are considered investigational or cosmetic. Providers are encouraged to inform members before rendering such services that the members are likely to be financially responsible for the cost of these services.

## DESCRIPTION

The vertical expandable prosthetic titanium rib (VEPTR) is a curved rod placed horizontally in the chest that helps to shape the thoracic cavity in children with spinal and thoracic deformities.

## MEDICAL POLICY CRITERIA

I. Use of the Vertical Expandable Prosthetic Titanium Rib may be considered **medically necessary** in the treatment of progressive thoracic insufficiency syndrome due to rib and/or chest wall defects in infants/children between six months of age and skeletal maturity.

II. Use of the Vertical Expandable Prosthetic Titanium Rib for all other conditions, including but not limited to the treatment of scoliosis in patients without thoracic insufficiency, is considered **investigational**.

*NOTE: A summary of the supporting rationale for the policy criteria is at the end of the policy.*

## POLICY GUIDELINES

It is critical that the list of information below is submitted for review to determine if the policy criteria are met. If any of these items are not submitted, it could impact our review and decision outcome.

- History and Physical/Chart Notes
- Documentation of syndrome and evidence of cause including age and skeletal maturity

## CROSS REFERENCES

None

## BACKGROUND

The vertical expandable prosthetic titanium rib (VEPTR) is a curved rod placed vertically in the chest that helps stabilize and shape the thoracic cavity. It is positioned either between the ribs or between the ribs and either the spine or pelvis. VEPTR may be described as a “rib-based” growth sparing instrumentation, which is compared with “spine-based” growing rods for Cobb angle correction. It is being evaluated for use in skeletally immature patients with thoracic insufficiency syndrome (TIS) and to slow or correct curve progression in pediatric scoliosis patients without TIS. The device is designed to be expanded every four to six months as growth occurs, and also to be replaced if necessary. Some patients require multiple devices.

TIS is the inability of the thorax to support normal respiration or lung growth.[1] It results from serious defects affecting the ribs or chest wall such as severe scoliosis, rib fusion (which may accompany scoliosis), and various hypoplastic thorax syndromes such as Jeune’s Syndrome and Jarcho-Levin syndrome. Spine, chest, and lung growth are interdependent. While the coexistence of chest wall and spinal deformity is well documented, their effect on lung growth is not completely understood.

Progressive TIS includes respiratory insufficiency, loss of chest wall mobility, worsening three-dimensional thoracic deformity, and/or worsening pulmonary function tests. As a child grows, progressive thoracic deformity and rotation toward the concave side occurs with worsening respiratory compromise. This progression is often accompanied by a need for supplemental oxygen and can require mechanical ventilation. While spinal fusion is one approach to treatment, it may not be successful and may also limit growth (lengthening) of the spine.

Given the complexity of these procedures and patients, implantation of this device should be performed in specialized centers. Preoperative evaluation requires input from pediatric orthopaedists, pulmonologist, and thoracic surgeon. In addition, preoperative evaluation of nutritional, cardiac and pulmonary function (when possible) is required.

### REGULATORY STATUS

The Vertical Expandable Prosthetic Titanium Rib (VEPTR) (DePuy Synthes Spine) device received US Food and Drug Administration (FDA) approval under a humanitarian device exemption (HDE) for the treatment of TIS in skeletally immature patients. The FDA review defined TIS as, “the inability of the thorax to support normal respiration or lung growth” and created the following categories to aid in the identification of potential TIS patients:

- Flail chest syndrome
- Rib fusion and scoliosis

- Hypoplastic thorax syndrome, including:
    - Jeune's syndrome
    - Achondroplasia
    - Jarcho-Levin syndrome
    - Ellis van Creveld syndrome

The FDA review noted that the device should not be used in patients less than six months of age. Skeletal maturity occurs at about age 14 for girls and age 16 for boys.

In 2014, the FDA cleared the Vertical Expandable Prosthetic Titanium Rib (VEPTR OR VEPTR II) through the 510(k) process. The device is indicated for skeletally immature patients with severe, progressive spinal deformities and/or three dimensional deformity of the thorax associated with or at risk of TIS (defined above). This would include patients with progressive congenital, neuromuscular, idiopathic, or syndromic scoliosis.

The predicate devices listed in the VEPTR 510(k) approval were the Medtronic CD HORIZON® Growth Rod Conversion Set and the Medos Sarl ISOLA® and EXPEDIUM® Growing Spine Systems.

## EVIDENCE SUMMARY

The most clinically relevant outcomes include the following:

- The ability of the device to restore and maintain a more normal anatomical shape while allowing for the continued growth of the child until skeletal maturity
- Durability of any beneficial treatment effects
- Safety, including the rate of adverse effects and reoperations

The following is a summary of the current published evidence for the use of VEPTR in the treatment of thoracic insufficiency syndrome (TIS) and scoliosis without TIS.

### THORACIC INSUFFICIENCY SYNDROME

The current evidence for the use of VEPTR in the treatment of TIS is limited to data from case series and retrospective reviews.[2-12] Results from these case series have consistently demonstrated improvement and/or stabilization in key measures with the use of VEPTR in children with TIS, with or without congenital scoliosis. This improvement was noted in measures related to thoracic structure, growth of the thoracic spine and lung volumes, and stable or improved ventilatory status.

In general, conclusions based on data from small case series are uncertain, largely due to the lack of an appropriate comparison group. However, since TIS is a rare disease with limited treatment options, it is unlikely that data from large RCTs will become available. In addition, TIS is a progressive disease with a natural history of worsening deformity, pulmonary function, and pulmonary insufficiency that is unlikely to improve in the absence of intervention. Therefore, the available case series evidence, which consistently reports improved health outcomes, is considered sufficient to conclude that VEPTR may provide a treatment benefit in select patients.

### SCOLIOSIS WITHOUT THORACIC INSUFFICIENCY SYNDROME

A study by Chen (2017) compared VEPTR to spinal growing rods (GR), with a focus on sagittal profile deformity (as opposed to the commonly studies coronal plane deformity).[13] This retrospective case series included 11 patients who had VEPTR, and 22 patients treated with GR. Similar coronal correction and spinal growth results were seen for both treatments, but GR appeared to be more effective in controlling thoracic kyphosis. The authors also noted higher overall complication rate with VEPTR (72.7%) than with GR (54.5%).

El-Hawary (2017) published a prospective multi-center observational cohort study that evaluated VEPTR insertion outcomes.[14] The review focused on spine growth and scoliosis progression data from 63 patients preimplant, post implant and at a two-year follow-up. At the two-year follow-up, VEPTR effectively treated patients with early onset scoliosis, without rib abnormalities. Eighty-six percent of the patients had improved scoliosis and 94% had satisfactory spine growth.

Livingston (2015) published results form a retrospective multicenter national database study that compared the efficacy of VEPTR versus GR patients with neuromuscular scoliosis who had a follow-up for greater than one year.[15]Twenty-three patients treated with VEPTR and 22 patients with GR were included in the study. The parasol score (T6 convex hemithoracic width/T6 concave hemithoracic width) × (T6 thoracic width/T12 thoracic width) and the assisted ventilation rate (AVR) did not improve after treatment with either the VEPTR or GR.

Farley (2014) compared treatment of congenital scoliosis with VEPTR (n=22) to treatment with spinal fusion (n=27) or observation (n=184) based on a prospective, consecutive, nonrandomized registry.[16] Patients with non-congenital scoliosis (idiopathic or neuromuscular) or with other vertebral disorders were excluded. Outcomes were measured with the Scoliosis Research Society (SRS-22) questionnaire which includes six domains: total, function, mental health, image, satisfaction, and pain. Compared to the observation group, the VEPTR group had higher total scores and image scores in the second and third visits and higher function scores between the third and fourth visits. A comparison between the fusion and VEPTR groups was not reported. Interpretation of this study is limited due to a number of confounding factors, including age at treatment, unknown comorbidities, and the rationale for the selection of treatment, and baseline questionnaires completed by the parents of younger children. In addition, comorbidities were not included in the study. These outcomes need to be validated in further prospective studies.

White (2011) reported the off-label use of spine-to-spine VEPTR to treat spinal deformity in 14 children without chest wall abnormalities.[17] The indications for the dual spine-to-spine rods were absence of a primary chest wall deformity, progression of spinal deformity to a Cobb angle of greater than 50 degrees, and migration of a previously placed proximal rib anchor or of a prior non-VEPTR growing rod to the point of loss of stable fixation. At final follow-up (24 to 48 months), there was an improvement in the Cobb angle from 74 to 57 degrees, an increase in T1-S1 height from 260 to 296 mm, and no significant change in kyphosis. Complications occurred in six of 14 patients (43%) and included three rod fractures in two patients, three superficial infections, and one case of prominent hardware that threatened skin integrity. The authors concluded that while results are similar to those obtained with other growing rods, “the high complication rates, need for multiple procedures in growing children, and small relative gains in radiographic parameters still challenge proof of efficacy of all such treatment methods.” Additional trials with more patients and longer duration are needed before conclusions about the safety and effectiveness of the VEPTR device in pediatric patients with spinal deformity, but without TIS, can be made.

## ADVERSE EFFECTS

A study published in 2017 evaluated 539 records for nerve injury alerts and actual nerve injuries during VEPTR implant, revision, expansion and/or removal. Three injuries occurred during implant, but the patients fully recovered. There were no reported injuries during expansion, revision, or removal. Level of evidence IV-diagnostic study.

One 2016 report concluded device related complications occurred in 22 out of 65 patients treated at a single center for TIS over a 13-year period. Complications were more likely in those with normal or hyperkyphotic curves. The majority of complications were managed by an additional surgical procedure.[18]

Complications associated with this device need to be considered by practitioners and families. Data from the FDA review and the papers by Campbell and Emans[2-4] reported a 25% device migration rate (although no significant long-term consequences were associated with this complication), a 10% rate of infection-related complications, and a 1% to 7% rate of brachial plexus injury or thoracic outlet syndrome.

Other adverse effects that have been reported include:[6,17,19-23]

- Implant breakage
- Rib fractures
- Hip joint destabilization
- Perforation of the iliac ala
- Seroma
- Spontaneous ossification (e.g., lumbar spine, rib, iliac crest)
- Tissue reaction to metal
- Esophageal rupture
- Infection

## PRACTICE GUIDELINE SUMMARY

No evidence-based clinical practice guidelines were identified which specifically recommend the use of the VEPTR for treatment of TIS or scoliosis in the absence of TIS.

## SUMMARY

There is enough research to show that the Vertical Expandable Prosthetic Titanium Rib (VETPR) may improve health outcomes for progressive thoracic insufficiency syndrome (TIS) due to rib and/or chest wall defects in infants/children between six months of age and skeletal maturity. This is a rare disorder with limited treatment alternatives. Therefore, use of the VEPTR device may be considered medically necessary in skeletally immature children at least six months of age with progressive thoracic insufficiency syndrome due to rib and/or chest wall defects.

There is not enough research to show that Vertical Expandable Prosthetic Titanium Rib (VETPR) improves health outcomes for people with any other conditions, including but not limited to the treatment of scoliosis in patients without thoracic insufficiency. No clinical guidelines based on research recommend VEPTR for people with any other conditions.

Therefore, use of VEPTR for the treatment of all other indications, including but not limited to the treatment of scoliosis in patients without thoracic insufficiency is considered investigational.

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## CODES

| Codes | Number | Description |
|---|---|---|
| CPT | 20999 | Unlisted procedure, musculoskeletal system, general |
| | 21899 | Unlisted procedure, neck or thorax |
| | 22899 | Unlisted procedure, spine |
| HCPCS | None | |

***Date of Origin:*** *June 2007*